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Rare case of cephalopagus conjoined twin with postmortem MRI

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Abstract

Conjoined twins are a rare and extreme form of monozygotic twinning. The frequency of conjoined twins is approximately 1 in 50,000 gestations, but many die in utero, are terminated or stillborn. Imaging plays a crucial role in the antenatal and postnatal life of conjoined twins, with the initial modality for screening being ultrasound followed by other modalities such as computed tomography (CT) and/ or magnetic resonance imaging (MRI).

We report a case of cephalopagus conjoined twins of 19 weeks 5 days of gestation. Ultrasound showed conjoined twins fused from head to the umbilicus with a single face and head, two posterior fossas, two hearts (unequal sizes) and eight limbs. MRI and X-ray of the abortus were performed to determine detailed anatomy of internal conjoined structures.

Antenatal imaging by ultrasound plays a vital role in the early intrauterine diagnosis of conjoined twins. An early prenatal diagnosis and investigation of the organs is very important for counselling the parents about the termination or continuation of the pregnancy and about the potential delivery and possible surgical solution.

Keywords: MRI, CT, Ultrasound

Introduction

Conjoined twins are a rare and extreme form of monozygotic twinning. The frequency of conjoined twins is approximately 1 in 50,000 gestations, but many die in utero, are terminated or stillborn. The true incidence is estimated to be around 1 in 250,000 live births. Like all monozygotic twins, all conjoined twins have the same sex. There is a distinct female predominance with a 3:2 female to male ratio [1]. Conjoined twinning is a random event, unrelated to heredity, maternal age or parity [2].

Imaging plays a crucial role in the antenatal and postnatal life of conjoined twins, with the initial modality for screening being ultrasound followed by other modalities such as computed tomography (CT) and/ or magnetic resonance imaging (MRI) [3].

They are classified based on the site of fusion. Two bodies can be fused in a symmetrical or asymmetrical alignment. The symmetrical alignments are side-by-side union or facing fusion. In the case of facing union, both co-twins share the conjoined portion; however, in the case of side-by-side union, a dominant co-twin may be present. Conjoined twins show varying degrees of conjoining [4]. The main types are omphalopagus

(abdomen), thoracopagus (thorax), cephalopagus (ventrally—head to umbilicus), is chipagus (pelvis), parapagus (laterally—body side), craniopagus (head), pygopagus (sacrum), and rachipagus (vertebral column) [5].

Case report

We report a case of cephalopagus conjoined twins of 19 weeks 5 days of gestation. A 26-year-old female (G4P2L2A1) with 4 months of amenorrhea was found to have conjoined twins on routine ultrasound and was referred to our institution for further evaluation. Ultrasound showed conjoined twins fused from head to the umbilicus with a single face and head, two posterior fossas, two hearts (unequal sizes) and eight limbs.

Decision for medical termination of pregnancy was taken in consultation with department of pediatric surgery. Procedure was uneventful with no complication and babies were still born. Postmortem examination was refused by the parents however consent for post-natal MRI and X-ray was given.

MRI and X-ray of the abortus were performed to determine detailed anatomy of internal conjoined structures.

Post-natal X-ray showed fused single cranium, single face, two spines and eight limbs (Fig. 2). MRI showed single large cranium with single cerebral hemisphere with multilayered appearance, smooth surface, notch of sylvian fissure and relatively large ventricles which appeared to be normal for age. There were two posterior cranial fossa containing two pons, medulla and cerebellum which continued below into two spinal cords through separate foramen magnum.

At the level of thorax, there were two hearts with more developed and larger heart anteriorly. Thoracic aorta of the left fetus was in continuation with the anteriorly placed heart. Thoracic and suprarenal aorta of right sided fetus was not well appreciated in MRI. There was a single trachea and an esophagus with four lungs. In abdomen, anteriorly placed liver appeared large and well formed with left and right lobe. The falciform ligament and umbilical vein from a single cord were terminating into it. Posterior liver was relatively small with both livers fusing, with a small slip-on right side. There were two kidneys on right and left side with fused adrenal gland on right side. Bowel loops were appreciated in the Centre of abdomen.

Discussion

Conjoined twins are born because the non-separated parts of the otherwise normal twins remain fused throughout the period of development. 70-80% cases of conjoined twins are females. Approximately 18-40% cases are joined at the chest (thoracopagus), 10-34% at the anterior abdominal wall (xiphopagous or omphalopagus), 18% at the buttocks (pygopagus), 6% at the ischium (is chiopagus), and 2-6% at the head (craniopagus). Forty percent of conjoined twins are still born, and an additional 35% survive only one day [6].

The exact cause of cephalopagus has still not been explained. In such cases, defects of blast oogenesis during the midline formation and other multiple mechanisms of abnormal embryogenesis have been proposed as being responsible for the abnormal development. It has been suggested that the initial fission of the notochordal anlage (and the primitive streak and primitive pit) with the subsequent formation of two centres of growth and fusion of the developing embryos could give rise to conjoined twins [4].

The attachment site as well as the extent of attachment determines the prognosis in conjoined twins. Due to the near infinite variations, the outcome is highly case-

specific. Prenatal diagnosis is possible using ultrasound, MRI, and CT, and can be performed as early as 12 weeks of gestation. Due to the presence of several congenital anomalies associated with conjoined twins, preoperative, intraoperative, and postoperative management is essential to improve prognosis. Cardiovascular and respiratory failures are the two major risk factors for death of the twins post-separation. There are many ethical challenges associated with treating conjoined twins, such as difficult decisions regarding early termination and determining the individuality of each twin.

The incidence of conjoined twins has declined after the advent of ultrasound guided maternal screening. We have presented this case owing to the rarity of the case and to emphasize the importance of timely antenatal screening.

Conclusion

Antenatal imaging by ultrasound plays a vital role in the early intrauterine diagnosis of conjoined twins. The prognosis for conjoined twins is dependent on the associated anomalies present and on the degree of fusion of the intra-cranial, intra-thoracic and/or intra-abdominal structures. An early prenatal diagnosis and investigation of the organs is very important for counselling the parents about the termination or continuation of the pregnancy and about the potential delivery and possible surgical solution. In developing countries, like India, where there are limitations in expertise and technology, detailed counseling of the parents is required to explain long-term outcomes. When termination of pregnancy is opted, it should be done at an early stage as later stages are fraught with problems.

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Legend Figures

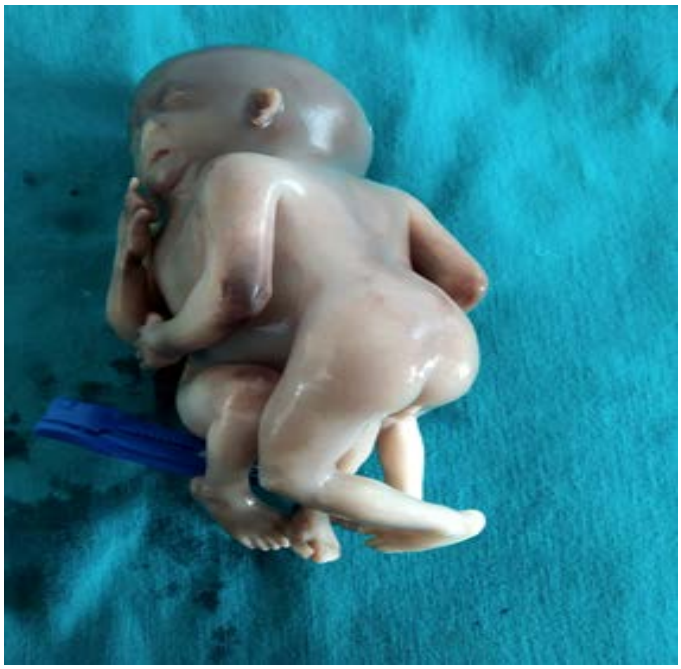

Fig 1: Gross image of the abortus.

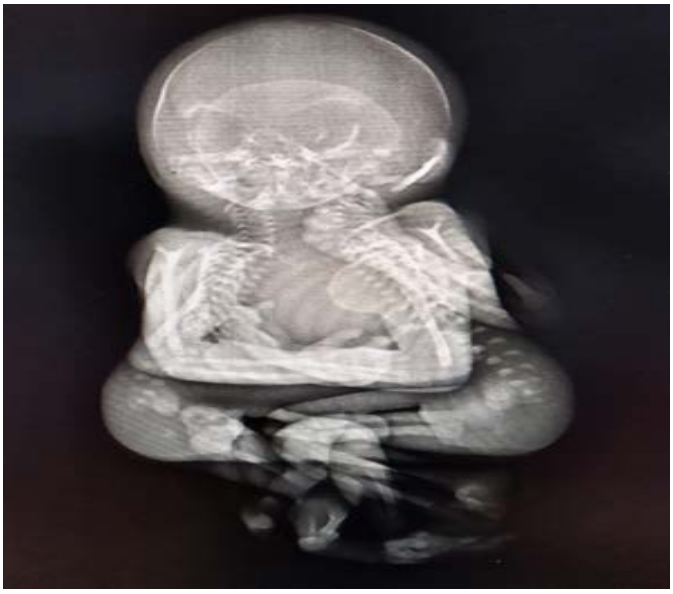

Fig 2: X-ray showing fused single cranium, single face, two spines and eight limbs.

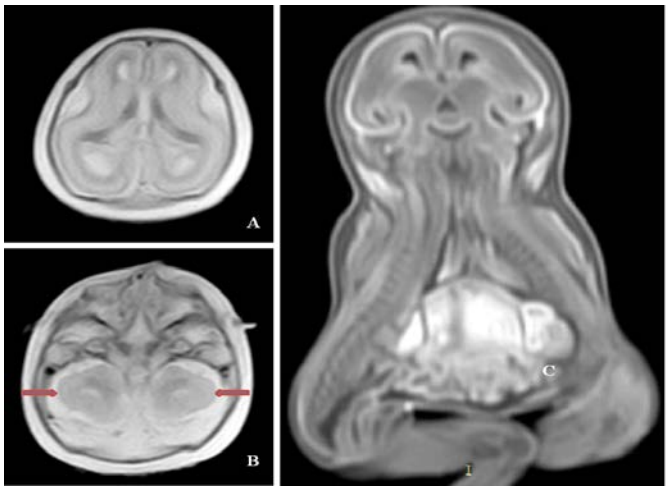

Fig 3: T2WI axial MRI images (A, B) showing single cerebrum and two cerebellum. Coronal T1 FS image (C) showing two spinal cords passing through separate foramen magnum

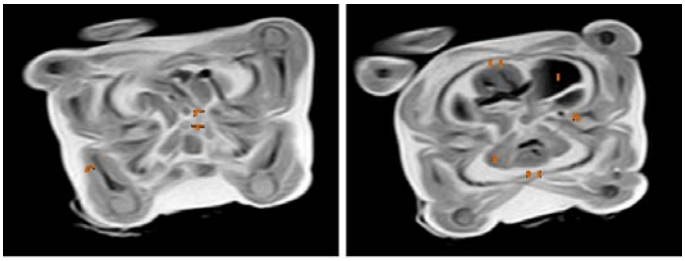

Fig 4: Axial T2WI MRI of upper and lower thoracic cavity showing different structures. S: shoulder, E: esophagus, T: trachea, H1and H2: heart , L: lung , A: aorta of left fetus, L: liver.

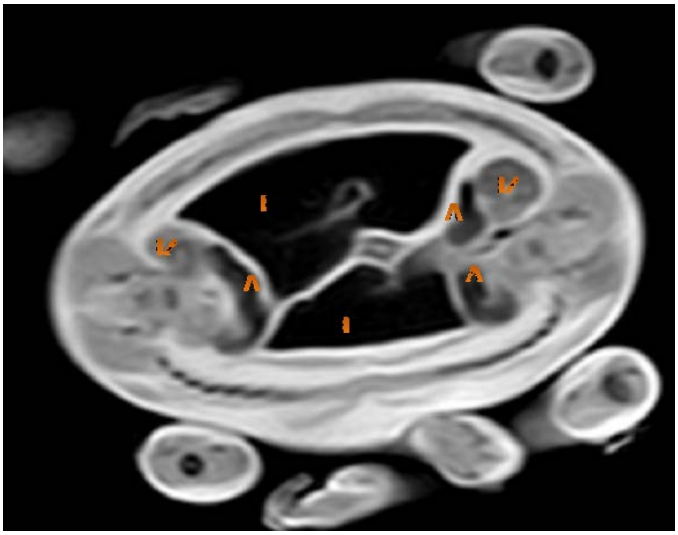

Fig 5: Axial T2 WI of upper abdomen showing fused adrenal glands (A1) of right fetus and normal on left side (A2). Liver (L) on anterior aspect is well formed. K: kidney.